Movement Disorders
CLINICAL PRACTICE

CASE REPORT

Photosensitive Epilepsy Syndromes Mimicking Motor Tics

Katherine Longardner, MD,[1,*] Soaham Desai, MD, DM,[2,3] Mark Nespeca, MD,[1,4,5] Matthew Bainbridge, PhD,[6] and Jennifer Friedman, MD[1,4,5,6]

About 3% of children are clinically diagnosed with tics,[1] although at least 1 observational study suggests that the prevalence may be much higher—up to 25%.[2] Motor tics may manifest as simple movements (eg, blinking, head jerking) or a complex series of movements. Stereotypies are even more common, with estimated prevalence in children ranging from 50% to >90%.[3] Given the wide range of manifestations and body parts affected, the differential diagnosis for these childhood movement disorders is broad. Similar to tics and stereotypies, seizures may manifest as sudden onset, episodic, repetitive, rhythmic movements. In this report of 2 children with rare photosensitive epilepsies mimicking tic-like movements, we illustrate the overlapping phenomenology of seizures and common pediatric movement disorders. Jeavons syndrome (case 1) is characterized by eyelid myoclonia with or without altered awareness, eye closure–induced generalized paroxysms, and photosensitivity.[4] Sunflower syndrome (case 2) is named for the heliotropic flower, as patients appear drawn to the sun or other bright light during seizures.[5,6] Affected patients often stare at light while repetitively waving their hand in front of their eyes; the flickering light created by this movement may induce the seizures.[7] Differentiating tics and stereotypies from seizures is essential for proper treatment and prognostic counseling. We discuss clues that can help distinguish seizures from childhood movement disorders sharing similar phenomenology.

Case Report

Case 1

A 6-year-old boy with normal birth and development was referred for suspected tics, with blinking episodes occurring 15 to 20 times daily. Examination was normal. Because no abnormal movements were observed, a home video of the episodes was reviewed (Video S1). The video showed eyelid closure–induced rapid blinking associated with upward gaze deviation suggestive of eyelid myoclonia. The patient also had altered awareness and photosensitivity when looking toward the light from the window. These clinical features suggested photosensitive seizures, which prompted further evaluation for epilepsy. An electroencephalography (EEG) revealed bursts of generalized sharp and slow wave discharges associated with eye closure, photoparoxysmal response on photic stimulation, and intermittent generalized polyspike and wave discharges (Fig. 1). Given the clinico-electrographic findings, he was diagnosed with Jeavons syndrome. His seizures resolved with valproic acid 15 mg/kg/day. The patient's father recalled that during his childhood, others observed him having similar eyelid movements while playing outdoors. The father was later diagnosed with epilepsy after experiencing a generalized seizure at age 17 years; his epilepsy was also controlled on valproic acid.

Case 2

An 8-year-old normally developing boy presented for evaluation of abnormal movements beginning at age 6 years. The most frequent movement was waving his left hand in front of his face, which occurred multiple times daily and was distractible and suppressible. There was no premonitory urge. His mother reported that he appeared "spaced out" during the movements, but could answer questions and respond appropriately while hand waving. Movements occurred most frequently when outdoors, especially while playing baseball, but did not impede his athletic performance, as shown in a home video (Video S2). Multiple other movements and vocalizations were endorsed. Family history was notable only for generalized epilepsy in his mother since age 19 years. Examination was normal.

[1]Department of Neurosciences, University of California San Diego, San Diego, California, USA; [2]Pramukhswami Medical College, Karamsad, India; [3]Shree Krishna Hospital, Gokal Nagar, India; [4]Department of Pediatrics, University of California San Diego, San Diego, California, USA; [5]Division of Neurology, Rady Children's Hospital San Diego, San Diego, California, USA; [6]Rady Children's Institute of Genomic Medicine, San Diego, California, USA

***Correspondence to:** Dr. Katherine Longardner, Department of Neurosciences, University of California San Diego, 9500 Gilman Dr., Mail Code 0886 La Jolla, CA 92023; E-mail: klongardner@ucsd.edu
Keywords: Tourette syndrome, tics, stereotypies, pediatrics, epilepsy.
Katherine Longardner and Soaham Desai contributed equally to this work.
Relevant disclosures and conflicts of interest are listed at the end of this article.
Received 7 March 2020; revised 3 May 2020; accepted 18 May 2020.
Published online 00 Month 2020 in Wiley Online Library (wileyonlinelibrary.com). DOI: 10.1002/mdc3.13053

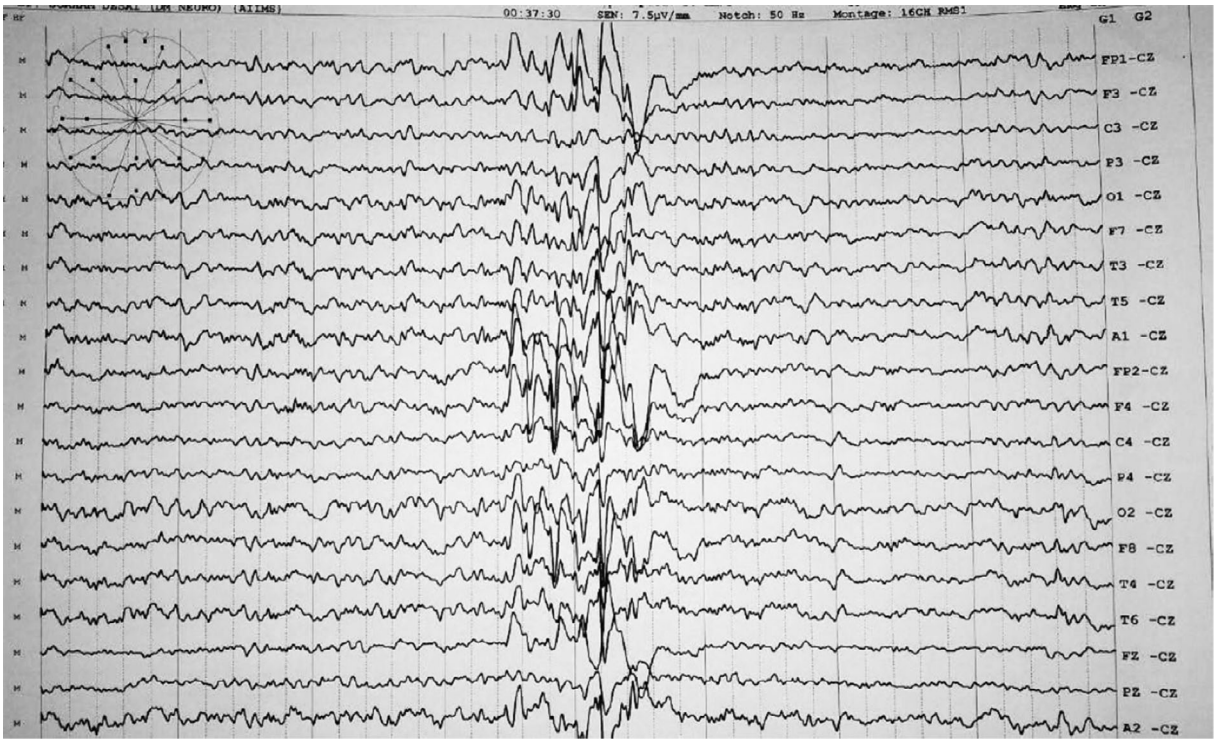

FIG 1. Electroencephalography showing generalized sharp and slow wave discharges associated with eyelid closure, confirming Jeavons syndrome.

A diagnosis of Tourette syndrome was made based on clinical history. However, the character of hand-waving movements observed on the video and the maternal history of epilepsy raised suspicion that the hand-waving movements might be seizures. Sleep-deprived EEG showed recurrent irregular generalized and multifocal independent epileptiform discharges during wake and sleep states. Photic stimulation did not produce any clinical event despite the presence of a photo-paroxysmal response. A video EEG was performed outdoors on a sunny day in an effort to document the reported finding of hand waving that occurred in sunlight, confirming typical prominent long-duration sunflower-type seizures (Video S3). The patient was advised to wear sunglasses when outdoors and was treated with valproic acid 250 mg twice a day, resulting in a reduction of sunflower seizures by approximately 50% to 75%.

On further questioning, the patient's mother revealed that she had photosensitive epilepsy. Given the family history and suspected genetic contribution to photosensitive epilepsy syndromes,[8] genetic testing was performed, but no clear causative abnormality was demonstrated (Appendix 1).

Discussion

As these cases illustrate, photosensitive epilepsies may mimic tics or stereotypies, which also begin during childhood and are characterized by repetitive, stereotyped movements that may be suppressible. Typically seizures are not suppressible, and following commands is not possible during generalized events. However, photosensitive seizures may have atypical features,[9] including possible volitional induction, or as demonstrated in case 2, preserved awareness. These unusual aspects that are more typical of tics and stereotypies may lead to misdiagnosis. Table 1 lists other epilepsy syndromes that may clinically resemble movement disorders.

Jeavons first described the classic tetrad of eyelid myoclonia with or without absences, eyelid-closure–induced EEG showing paroxysms and polyspike and wave discharges, photoparoxysmal response, and photosensitivity.[15] The prevalence of Jeavons syndrome is estimated between 7% to 13% of all generalized idiopathic epilepsies, but may be higher given the underrecognition of this uncommon epilepsy phenotype.[16] Eyelid myoclonia is an ictal phenomenon characterized by rapid, jerky blinking immediately after eye closure associated with mild upward gaze deviation.[16] Eyelid myoclonia may occur in other seizure disorders, including sunflower syndrome, juvenile myoclonic epilepsy, childhood absence epilepsy, and juvenile absence epilepsy.[15] This epileptic eyelid fluttering may appear similar to blinking motor tics (Table 1). However, eyelid myoclonia can be differentiated from tics by the rapid frequency of blinking, transient unresponsiveness, and photosensitivity. Although clarifying exact event semiologies often requires adept video camera angle

TABLE 1 Epilepsies that may mimic tics or other movement disorders

Seizure Semiology	Examples	Movement disorder mimics
Eyelid myoclonus with possible self-induction[10]	Jeavons syndrome, sunflower syndrome	*Blinking tics or hand-waving tics*
Absence seizures[10]	Childhood absence epilepsy, juvenile absence epilepsy, progressive myoclonic epilepsies	*Blinking tics*
Focal asymmetric myoclonus[10]	Facial myoclonias with absences	*Facial tics*
Sleep-related hypermotor epilepsy[11]	Autosomal dominant nocturnal frontal lobe epilepsy	*Paroxysmal hypnogenic dyskinesia (as seen in ADCY5 mutation carriers[12]) or parasomnias*
Tonic versive phenomenon[13]	Temporal lobe epilepsy	*Paroxysmal dystonia or tics*
Faciobrachial dystonic seizures[14]	Leucine-rich glioma-inactivated 1 (LGI1) antibody limbic encephalitis	*Paroxysmal dystonia or tics*

capture of the events in question, a general helpful rule is that upward vertical elevation of the eyes is common in eyelid myoclonias with absence including with Jeavons syndrome, whereas in typical childhood or juvenile absence epilepsy, the eyes usually are straight without movement above or below a horizontal meridian. However, the presence or absence of upward eye deviation is not helpful in distinguishing tics from epileptic phenomena.

Sunflower syndrome is another rare photosensitive epilepsy that usually manifests with eyelid myoclonia or focal seizures with impaired awareness. Seizure induction may be voluntary to produce a self-soothing or self-gratifying stimulus. However, it is incompletely understood whether hand waving causes flickering light that induces seizures or whether the movement is a part of the seizure itself.[7] Our patient denied an urge or pleasurable feeling associated with hand waving and noted that he could suppress the movements. Sunflower syndrome can be differentiated clinically from tics or stereotypies by photosensitivity, that is, always occurring while facing bright light. In our case, the patient's retained awareness and the distractibility of the behavior as well as parental endorsement of multiple movements and vocalizations was misleading. This patient likely has both sunflower syndrome and Tourette syndrome given the clinical history of numerous vocalizations and movements in addition to the stereotyped hand waving associated with generalized polyspike and wave discharges on EEG.

Most patients with Jeavons syndrome respond well to valproic acid monotherapy, however, 30% to 40% require adjunctive antiepileptic treatment, including levetiracetam, topiramate, or lamotrigine. Valproic acid has also been used with success in sunflower syndrome, although the sample sizes are small.[17] Because these epilepsies typically require lifelong therapy, levetiracetam or lamotrigine may be preferred in affected women because of valproic acid's potential teratogenicity.[18] Both Jeavon's syndrome and sunflower syndrome belong to a subgroup of generalized photosensitive reflex epilepsies, which includes Pokémon seizures, among others.[19] Gene mutations have been identified in a subset of patients with photosensitive epilepsy syndromes, although the genetic etiology for the majority has not yet been ascertained.

Both patients had a parent with generalized epilepsy and photosensitive seizures, suggesting a genetic contribution. Case 1 did not undergo genetic testing, but pathogenic variants in several genes have been identified in individuals with Jeavons syndrome, including *KIAA2022* and *SLC2A1*.[20,21] Although a family history of photosensitive seizures and other epilepsy syndromes are common in photosensitive epilepsies, no specific gene has yet been identified as causative in sunflower syndrome.[8,22] Extensive genetic testing in case 2 was inconclusive. To our knowledge, although several chromosomal loci and possible susceptibility genes have been associated with individuals with photosensitive epilepsy,[8] no genetic association has previously been reported in an individual with sunflower syndrome. Further research will be needed to determine if other individuals with sunflower syndrome or other photosensitive epilepsies share variants in any of the genes identified in case 2.

Given the overlapping semiology with more common pediatric movement disorders and significant treatment implications, it is important for clinicians to include photosensitive epilepsies in the differential diagnosis of stereotyped movements in the appropriate clinical context (eg, occurring in bright light, associated with alteration of awareness, positive family history of epilepsy). Furthermore, tics and stereotypies are common in children and may coexist with seizure disorders. If epilepsy is considered, home video may be helpful, and EEG with time-locked video recording is essential to differentiate stereotyped seizures from nonepileptic movements.

Acknowledgments

We thank the patients and their families. We acknowledge support for this effort from Rady Children's Institute for Genomic Medicine.

Author Roles

(1) Research Project: A. Conception, B. Organization, C. Execution; (2) Manuscript Preparation: A. Writing of the First Draft, B. Review and Critique; (3) Interpretation of Genetic Findings; (4) Video Editing.

K.L.: 1A, 1B, 1C, 2A, 2B, 4
S.D.: 1A, 1B, 1C, 2A, 4
M.N.: 2B, 4
M.B.: 2B, 3
J.F.: 1B, 1C, 2B, 3, 4

Disclosures

Ethical Compliance Statement: In Case 1, the patient's mother provided informed consent for presentation and publication. Case 1 was approved by the institutional ethics committee. In Case 2, the patient's mother provided written informed consent to publish the patient's case and to publish videos, including facial images. The authors confirm that the approval of an institutional review board was not required for this work. We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines.

Funding Sources and Conflicts of Interest: No specific funding was received for this work. The authors declare that there are no conflicts of interest relevant to this work.

Financial Disclosures for the Previous 12 Months: Soaham Desai has no financial disclosures. Katherine Longardner served on the Acadia Fellows Advisory Board for Parkinson Disease Psychosis (2019). Mark Nespeca has received compensation from Zogenix, Inc for an investigation drug trial in Dravet syndrome. Matthew Bainbridge is the founder of Codified Genomics LLC and consults with Baebies Inc. Jennifer Friedman's spouse is founder and principal of Friedman Bioventure, which holds a variety of publicly traded and private biotechnology interests. In addition, he is chief operating officer of DTX Pharma, which is a company developing RNA therapeutics.

References

1. Knight T, Steeves T, Day L, Lowerison M, Jette N, Pringsheim T. Prevalence of tic disorders: a systematic review and meta-analysis. *Pediatr Neurol* 2012;47(2):77–90.
2. Snider LA, Seligman LD, Ketchen BR, et al. Tics and problem behaviors in schoolchildren: prevalence, characterization, and associations. *Pediatrics* 2002;110(2) Pt 1:331–336.
3. Muthugovindan D, Singer H. Motor stereotypy disorders. *Curr Opin Neurol* 2009;22(2):131–136.
4. Jeavons PM. Nosological problems of myoclonic epilepsies in childhood and adolescence. *Dev Med Child Neurol* 1977;19(1):3–8.
5. Bettoni L, Bortone E, Ghizzoni P, Juvarra G. The sunflower syndrome. *Funct Neurol* 1986;1(2):175–181.
6. Akbas Y, Oz Tuncer G, Serdaroglu A. A rare self-induced reflex epilepsy: sunflower syndrome. *Acta Neurol Belg* 2019;119(4):617–618.
7. Ames FR, Saffer D. The sunflower syndrome. A new look at "self-induced" photosensitive epilepsy. *J Neurol Sci* 1983;59(1):1–11.
8. Taylor I, Berkovic SF, Scheffer IE. Genetics of epilepsy syndromes in families with photosensitivity. *Neurology* 2013;80(14):1322–1329.
9. Xue J, Gong P, Yang H, et al. Genetic (idiopathic) epilepsy with photosensitive seizures includes features of both focal and generalized seizures. *Sci Rep* 2018;8(1):6254.
10. Covanis A. Photosensitivity in idiopathic generalized epilepsies. *Epilepsia* 2005;46((suppl 9)):67–72.
11. Tinuper P, Bisulli F. From nocturnal frontal lobe epilepsy to sleep-related hypermotor epilepsy: a 35-year diagnostic challenge. *Seizure* 2017;44:87–92.
12. Friedman JR, Meneret A, Chen DH, et al. ADCY5 mutation carriers display pleiotropic paroxysmal day and nighttime dyskinesias. *Mov Disord* 2016;31(1):147–148.
13. Kotagal P, Luders H, Morris HH, et al. Dystonic posturing in complex partial seizures of temporal lobe onset: a new lateralizing sign. *Neurology* 1989;39(2) Pt 1:196–201.
14. Irani SR, Michell AW, Lang B, et al. Faciobrachial dystonic seizures precede Lgi1 antibody limbic encephalitis. *Ann Neurol* 2011;69(5):892–900.
15. Striano S, Capovilla G, Sofia V, et al. Eyelid myoclonia with absences (Jeavons syndrome): a well-defined idiopathic generalized epilepsy syndrome or a spectrum of photosensitive conditions? *Epilepsia* 2009;50 ((suppl 5)):15–19.
16. Covanis A. Jeavons syndrome—updated review. *J Epileptol* 2015;23(2): 113–123.
17. Baumer FM, Porter BE. Clinical and electrographic features of sunflower syndrome. *Epilepsy Res* 2018;142:58–63.
18. Smith KM, Youssef PE, Wirrell EC, et al. Jeavons syndrome: clinical features and response to treatment. *Pediatr Neurol* 2018;86:46–51.
19. Verrotti A, Beccaria F, Fiori F, Montagnini A, Capovilla G. Photosensitivity: epidemiology, genetics, clinical manifestations, assessment, and management. *Epileptic Disord* 2012;14(4):349–362.
20. Madaan P, Jauhari P, Chakrabarty B, Gulati S. Jeavons syndrome in a family with GLUT1-deficiency syndrome. *Seizure* 2019;71:158–160.
21. Samanta D, Willis E. *KIAA2022*-related disorders can cause Jeavons (eyelid myoclonia with absence) syndrome. *Acta Neurol Belg* 2020;120(1): 205–207.
22. Karkare KD, Menon RN, Radhakrishnan A, Cherian A, Thomas SV. Electroclinical characteristics and syndromic associations of "eye-condition" related visual sensitive epilepsies-a cross-sectional study. *Seizure* 2018;58:62–71.

Supporting Information

Supporting information may be found in the online version of this article.

Appendix S1. Supplemental Data 1: Description of case 2 genetic testing. Supplementary Table 1: Identified variants in the proband. Gene, mutation, maternal inheritance, minor allele frequency, and associated disease with Mendelian Inheritance in Man ID and phenotype for each variant identified by sequencing lab.

Video S1. Home video showing 3 episodes of eyelid closure–induced rapid blinking associated with upward gaze deviation (seen in slow motion at the end of the video). This patient also had transient altered awareness and photosensitivity when looking toward the light while on the terrace or toward the window when indoors.

Video S2. Home video (the patient is the pitcher) showing stereotyped hand waving movements while looking at the sun, as demonstrated by his shadow. Movements did not interfere with baseball performance.

Video S3. Outdoor electroencephalography showing generalized polyspike and wave discharges time-locked with hand waving, confirming sunflower syndrome. During the hand waving and associated generalized epileptiform discharges, the patient was able to follow commands (eg, touch examiner's hand).